Journal of Surgical Oncology 2000;73:228–230

Benign Chondroid Syringoma: Report of a Case Clinically Mimicking a Malignant Neoplasm

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Chondroid syringoma, or mixed tumor of skin, is an uncommon sweat gland tumor most often seen in the head-and-neck region of patients in the sixth or seventh decade. Tumors usually present as asymptomatic, slowly growing masses. Histologically, there are both epithelial and stromal components. The treatment of choice is local excision. Rare malignant examples have been reported, commonly involving the extremities. We present a case of cutaneous chondroid syringoma arising in the thigh of a 28-year-old female. The tumor grew over a 4-year period, increasing rapidly in size over the last few months with fixation and pigmentation of the overlying skin clinically mimicking a malignant neoplasm. Such cutaneous appendage tumors are uncommon, and surgeons may be unfamiliar with them.
J. Surg. Oncol. 2000;73:228–230.

KEY WORDS: chondroid syringoma; mixed tumor of skin

INTRODUCTION

"Mixed tumor of the skin" was originally described by Billroth in 1859 as an entity histologically similar to the mixed tumor of the salivary glands. In 1961 Hirsch and Helwig [1] introduced the term "chondroid syringoma" for this sweat gland tumor [2,3]. Chondroid syringoma is a benign tumor characteristically presenting as an asymptomatic, slowly growing mass in the head-and-neck region of predominantly male, elderly patients. Malignant examples of chondroid syringomas have also been reported, and these most commonly present in the lower extremities of younger patients. Although the tumor is uncommon, we believe it is a neglected diagnosis which never appears in the differential diagnosis of the clinician. Herein, we present a case of benign chondroid syringoma, clinically mimicking a malignant neoplasm, involving the right thigh of a young female.

CASE REPORT

A 28-year-old woman presented with a 4-year history of a thigh mass. The mass originated as a slowly growing, painless, mobile, and soft to firm nodule with no attachment to the surrounding structures. Three years later, there was rapid growth of the nodule with increased pigmentation of the overlying skin. The nodule became hard, was fixed to the skin and surrounding structures, and measured about 2.5 cm in diameter. An excisional biopsy was performed based on the impression that the patient harbored a malignant neoplasm.

Grossly, there was a well-circumscribed, 2.5 cm, firm to hard nodule with a shiny, smooth, white cut surface. There was no necrosis or hemorrhage. Microscopic examination showed a dermal and subcutaneous neoplasm characterized by proliferation of clusters of epithelial cells with round to ovoid nuclei and, in some areas, forming tubular structures. The stroma had myxoid degeneration with areas of chondroid as well as osseous metaplasia (Figs. 1, 2). These features were in keeping with chondroid syringoma/pleomorphic adenoma/cutaneous mixed tumor of salivary gland type/mucinous hidradenoma. No significant pathological findings were seen in the overlying skin. Histologically, there were no malignant features such as vascular space invasion, increased or

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Accepted 7 January 2000

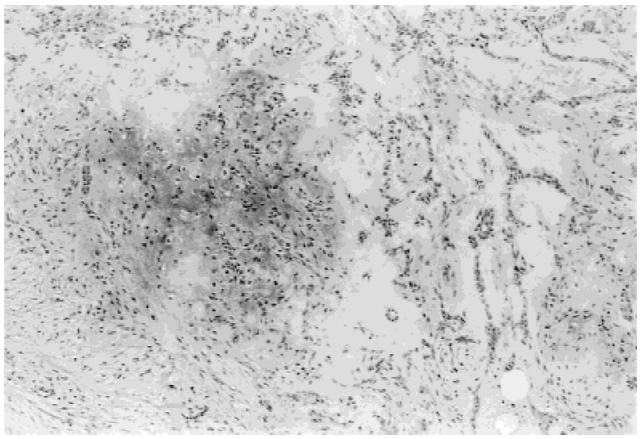

Fig. 1. Chondroid syringoma. Low magnification. Cords and tubules of epithelial cells proliferate in a chondroid background. Original magnification: 20×.

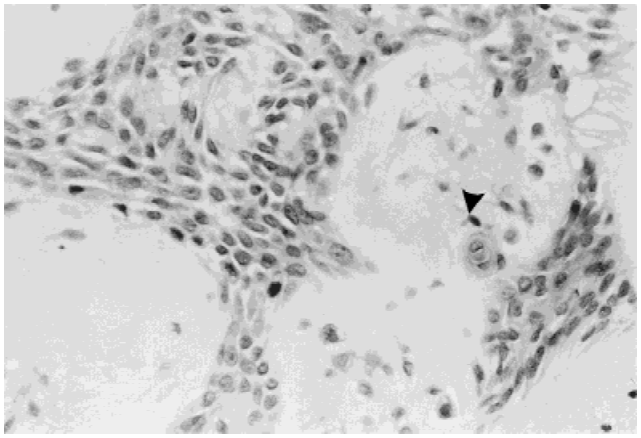

Fig. 2. Focal squamous differentiation with early squamous pearl formation (arrowhead) was seen in this tumor, presumably a reflection of its chronicity. Original magnification: 40×.

atypical mitoses, or cellular or nuclear pleomorphism. The patient remains free of recurrence or metastasis at 48 months.

DISCUSSION

Mixed tumors of the skin derive their name from the epithelial and mesenchymal elements seen on light microscopic examination. They feature cartilaginous metaplasia and adnexal structures [1,2]. Chondroid syringoma is an uncommon neoplasm, and most cases behave in a benign fashion, though rare malignant examples have been reported. Benign cutaneous mixed tumor occurs most commonly in the head-and-neck region, but other reported sites include the scrotum, eyelid, and brain [3–6]. The malignant variety most often involves the lower extremities of females [7].

Benign tumors usually present as asymptomatic, firm, subcutaneous or intracutaneous, slowly growing, solitary nodules. They are most frequently seen in middle-aged men. Malignant tumors, however, usually appear as firm, subcutaneous, usually slowly growing, single or multiple nodules; have no age predilection; and have a higher incidence in females [8]. They often arise in previously benign lesions; however, 1 case observed in the sacral area was associated with a peripheral low-grade malignant component of the adenocystic type [9].

Histologically, tubular or ductal structures, usually of eccrine or apocrine type, are set in a myxoid or chondroid matrix. Ossification of the stroma, and rarely sebaceous gland or hair matrix differentiation, may occur [10]. Nests of polygonal cells and keratinous cysts are commonly observed. Rarely hidrocystoma-like changes are seen in an otherwise typical chondroid syringoma [11]. Although scant amounts of a hyaline cell component can be seen, tumors with predominant or exclusive hyaline cell differentiation have been described, and these may pose diagnostic difficulty [12,13]. Malignant chondroid syringomas also consist of a combination of epithelial and mesenchymal elements but differ by the presence of polygonal epithelial cells with pleomorphism and increased mitotic counts of up to 5 mitotic figures per high-power field [14]. The epithelial cells form trabeculae, cords, islands, or tubules, though the latter is seen less frequently than in benign tumors. The stroma is almost always myxoid. The mesenchymal element is usually cartilaginous, and rarely ossification is present. The histological features that distinguish malignant from benign chondroid syringoma include cytological atypia, increased mitotic figures, and necrosis. Many malignant tumors have infiltrative margins or evidence of vascular space invasion. However, rare malignant tumors lack histological hallmarks of malignancy but still metastasize, reportedly as late as 17 years after the primary diagnosis [15]. This subgroup had abundant mucoid matrix and poor chondroid differentiation.

Immunohistochemically, the inner cell layer of the tubular structures of chondroid syringomas have distinct epithelial features and express cytokeratin (CK), carcinoembryonic antigen (CEA), and epithelial membrane antigen (EMA). The outer cell layer expresses vimentin, S-100 protein, neuron-specific enolase, and in some cases, glial fibrillary acidic protien (GFAP). These cells are negative for CK, CEA, EMA, and muscle-specific actin (MSA) [16]. Assessment of the immunohistochemical localization of intermediate filament proteins indicates coexpression of vimentin and K8.12 cytokeratin, as seen in pleomorphic adenoma of the salivary gland, suggesting that these outer tumor cells of the tubular structures have mixed properties of both epithelial and mesenchymal cells. The cells related to chondrogenesis display conspicuous S-100 protein and coexpress keratin and vimentin; thus, they are believed to arise from the modified myoepithelial cells of the tumor [17]. The lu-

minal epithelial cells bind the lectin *Ulex europaeus* and are CEA-positive [18].

Another immunohistochemical study of 9 cases of chondroid syringoma focused on the neoplastic myoepithelial cells, classifying them as hyaline (plasmacytoid), spindle, or polyhedral. Hyaline and polyhedral cells were negative for α-smooth muscle actin and MSA. CK14 stained a large number of polyhedral cells and only a small number of hyaline cells. Spindle cells were positive for CK14, smooth muscle actin and MSA. These findings strongly indicated that hyaline and spindle cells are of simple epithelial and myoepithelial types, respectively, whereas polyhedral cells show differentiation toward basal cells of the sweat gland ducts or myoepithelial cells [19].

To study the histogenesis of the apocrine type of chondroid syringoma, an immunohistochemical study using keratin expression was performed, which demonstrated that this type of tumor shows differentiation toward all components of apocrine units. Portions of keratinous cysts, the cuboidal luminal cells, and the columnar cells specifically differentiate toward the intrafollicular portion of the ducts and secretory cells of apocrine glands [20].

Electron microscopic findings establish the myoepithelial nature of the tumor cells; the ultrastructural features are similar to those of myoepithelial cells of pleomorphic adenomas (mixed tumors) of the salivary glands [21].

The features of benign chondroid syringomas are quite characteristic and usually pose little problem in histological diagnosis. The differential diagnosis of the malignant variety, however, includes carcinomas with mucinous stroma, mesenchymal neoplasms with chondroid differentiation, benign chondroid syringoma, and carcinosarcomas. However, close attention to the above-mentioned findings will aid in reaching the final diagnosis [22].

In summary, chondroid syringoma is an uncommon skin appendage tumor which almost never arises in a clinical differential diagnosis. The correct diagnosis is established by histological examination of the tumor. Our patient was a female with a slowly growing fixed mass with sudden accelerated growth in the thigh, clinically appearing as a malignant neoplasm and even histologically raising concerns about malignant chondroid syringoma, though lacking objective features of malignancy. Although the apparently benign histological features do not entirely exclude the possibility of subsequent malignant behavior, 48 months after the initial excision the patient remains alive and well without recurrence or metastasis.

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